

# Meningio-angiomas — case report and subject review

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## Introduction

Meningio-angiomas (MA) is a rare congenital hamartomatous malformation of the leptomeninges often also involving the adjacent cerebral cortex. Some cases are associated with neurofibromatosis (NF) whereas others develop in isolation.

## Case report

A 6-year-old girl presented after a prolonged grand mal seizure. She had suffered from recurrent focal seizures affecting her right side since 1 year of age and also suffered from a mild right-sided hemiplegia. There were no clinical or radiological stigmata of NF. An electro-encephalogram (EEG) demonstrated abnormal electrical activity over both temporoparietal regions.

An unenhanced computed tomography (CT) scan of the brain showed multiple areas of hyperintensity in the left parietal lobe extending in a linear radial fashion along a number of gyri and sulci over the surface of the brain (Fig. 1a). Moderate con-

trast enhancement was seen adjacent to some of these hyperintensities (Fig. 1b). The hyperintense areas appeared denser than blood. As there was no suspicion of any acute subarachnoid haemorrhage this was assumed to be calcification.

On unenhanced T1-weighted magnetic resonance (MR) images no direct evidence of any meningeal thickening could be seen, although

slight thickening and a mild increase in signal intensity was seen within the cortex over the convexity of the parietal lobe (Fig. 2a). No abnormal signal intensity was noted in this area on T2-weighted or proton-density images, although some prominent vessels were seen over the brain surface (Fig. 2b). Bright signal was, how-

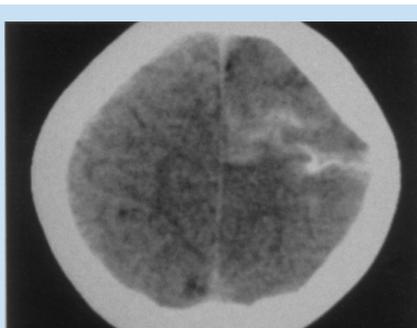


Fig 1a. Unenhanced axial CT scan showing linear hyperintense material over the surface of the left parietal lobe with extension into the sulci in keeping with calcified proliferative leptomeningeal tissue.

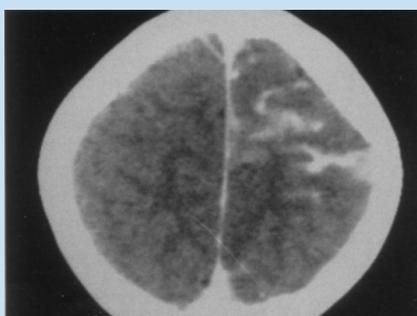


Fig 1b. Contrast-enhanced CT scan at the same level showing some areas of contrast enhancement within the abnormal tissue.

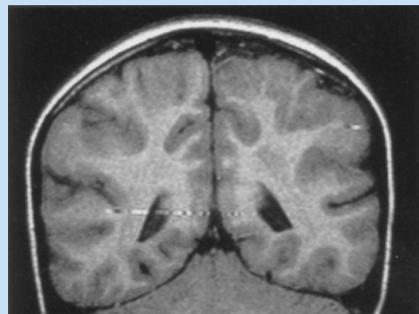


Fig 2a. Unenhanced coronal T1-weighted MR scan showing no meningeal thickening over the left parietal lobe but slight increase in the signal intensity of areas of the surface cortex itself.

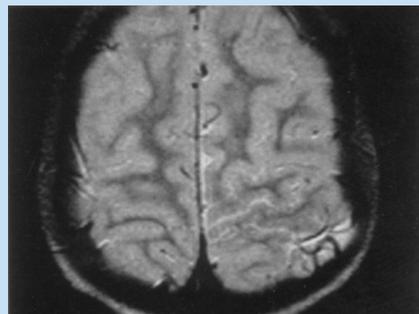


Fig 2b. No significant signal abnormality is noted on either standard T2-weighted or proton-density images (illustrated here). Some prominent surface vessels are seen.

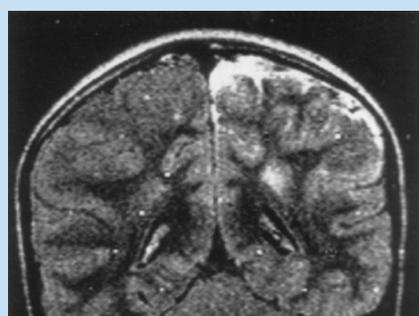


Fig 2c. Coronal FLAIR image showing bright signal arising from the thickened leptomeninges.

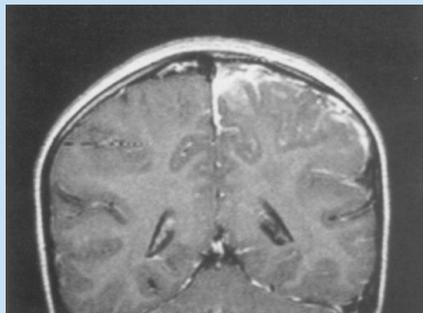


Fig 2d. Contrast-enhanced coronal T1-weighted image showing the enhancing thickened leptomeninges over the brain surface.

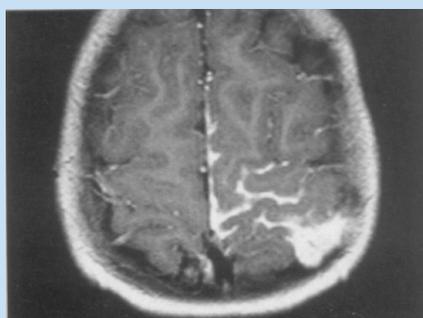


Fig 2e. Contrast-enhanced axial T1-weighted image at the same level as Fig 2b showing the densely-enhancing surface tissue and sulcal extension.

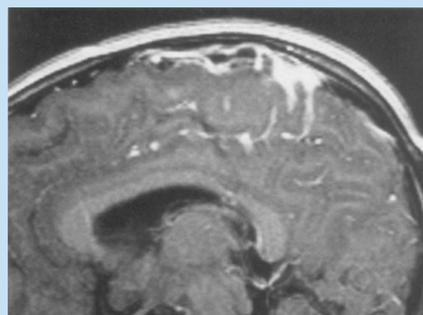


Fig 2f. Contrast-enhanced sagittal T1-weighted image showing the anteroposterior extent of the lesion. Note the radial extension within the sulci and possibly through the perivascular spaces as well.

ever, noted on the post-gadolinium FLAIR (Fig. 2c) and post-gadolinium T1-weighted images (Figs 2d - f).

Digital subtraction arteriography was then performed. Selective left external carotid arteriograms showed no abnormal dural supply, whereas selective left internal carotid arteriograms showed enlargement of the

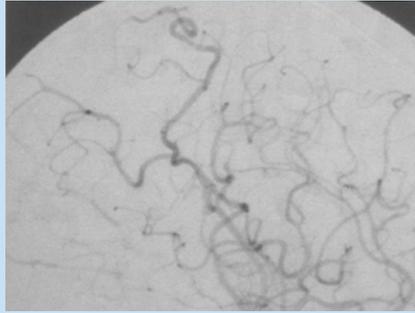


Fig 3a. Selective left internal carotid digital subtraction arteriogram, lateral projection, showing enlargement of the posterior parietal branch of the left middle cerebral artery corresponding to the prominent surface vessel seen in Fig. 2b.



Fig 3b. Same run, later image showing a local collection of slightly dysplastic looking branches over the affected cortex. No abnormalities were noted during the venous phase.

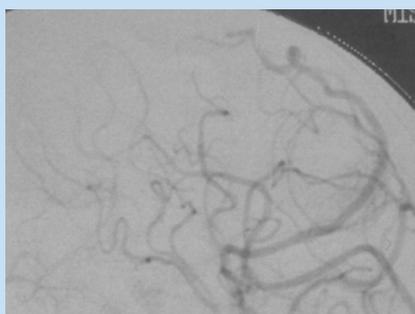


Fig 3c. Same vessel, right anterior oblique view, again showing the relatively prominent parietal branch.

posterior parietal branch of the left middle cerebral artery with some smaller slightly dysplastic looking branches seen in the same area as the abnormalities shown on the scan (Figs 3a - d).

The diagnosis of meningiomatosis was made on the basis of the clinical and radiological findings, but the

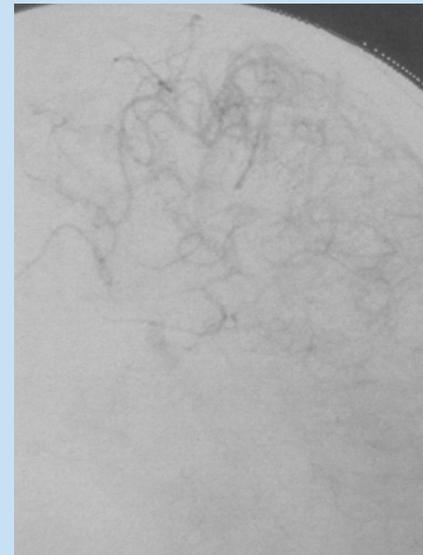


Fig 3d. Same run, later image again showing the slightly dysplastic-looking vessels over the cortex. No abnormalities of the external carotid dural supply were noted.

child has since been lost to follow-up with the result that no histological confirmation could be obtained.

## Discussion

MA is a rare benign hamartomatous lesion of the cerebral cortex and adjacent leptomeninges, first described by Bassoe and Nuzum<sup>1</sup> in 1915 in a patient with NF. The term 'meningio-angiomas' was first used in 1937 by Worcester-Drought et al.,<sup>2</sup> who also first suggested that MA may represent a forme fruste of NF. Many cases have been reported in NF patients, but not all are associated with NF.<sup>3-5</sup> In sporadic cases the male/female ratio is more equal and the mean patient age tends to be somewhat younger, children and young adults being affected. Patients typically present clinically with headaches and seizures but some cases are also found incidentally at autopsy<sup>6,7</sup> or during cranial imaging.<sup>8</sup> In most cases the MA affects the cerebral cortex, usually in the frontal or temporal regions. Some cases may show

involvement of the third ventricle, thalamus or brainstem.<sup>6,9</sup>

The characteristic pathological findings include leptomeningeal meningovascular fibroblastic proliferation and a variable degree of leptomeningeal calcification.<sup>3,10</sup> There is angiomatous proliferation within the meninges, sometimes with associated arteriovenous shunting. The fibroblastic and angiomatous proliferation can extend in a linear fashion along the Virchow-Robin perivascular spaces, thereby appearing to 'penetrate' the cortical grey matter.<sup>3</sup> The degree of calcification can vary from numerous psammoma bodies histologically to dense calcification and even ossification.<sup>8</sup> Changes may also be seen in the adjacent brain cortex (meningocephalo-angioneuromatosis). The cortical changes include the presence of neurofibrillary tangles, thought to represent degenerative changes in entrapped neurons.<sup>6</sup>

On unenhanced CT scans the calcifications may be seen as either linear or granular in nature,<sup>3</sup> whereas non-calcified areas or lesions may range from isodense to moderately hypodense with the low-density appearance probably representing a loosely packed cellular matrix consisting of

prominent perivascular spaces and scattered meningeal spindle (fibrous) cells.<sup>7</sup> The degree of contrast enhancement is variable. On MR scans the lesions tend to be iso- to hypointense to grey matter on T1-weighted images. Non-calcified areas or lesions are typically hyperintense on T2-weighted images with areas of dense calcification producing marked T2 shortening and thus appearing as hypo-intense areas often within the center of the lesion.<sup>3,8,10</sup> Again contrast enhancement is variable. Angiographic findings also vary from normal through the presence of an avascular 'mass' to the finding of abnormal vessels.<sup>4,8,10</sup>

The differential diagnosis of MA includes the Sturge-Weber syndrome, meningioma, granulomatous meningitis (sarcoid or tuberculosis) and calcified infiltrating glioma. MA can be readily differentiated from Sturge-Weber syndrome in that the calcification in the latter is parenchymal (gyral) and is associated with brain tissue atrophy on sectional imaging and a venous collateral ('pseudophlebatic') pattern in the affected regions at arteriography.<sup>3</sup>

Total surgical resection is the treatment of choice for MA, with the prog-

nosis being very good with complete cure from seizures in most cases.<sup>4-6</sup>

## References

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